

REPORT OF A CASE OF TUMOR OF THE CAROTID BODY.*

BY JOHN CHALMERS DA COSTA, M.D.,

OF PHILADELPHIA,

Professor of Principles of Surgery and Professor of Clinical Surgery in the Jefferson Medical College; Surgeon to the Philadelphia Hospital.

THE first person to suspect the existence of the carotid body seems to have been the great Haller; and from his time on there has now and then been a suggestion by some anatomist of a knowledge of the presence of the structure. In 1833 Mayer gave a description of it and pointed out its common situation in the bifurcation-angle of the carotid artery. He described it as about the size of a grain of rice, and as attached to the carotid vessel; and mentioned some other facts in relation to it. Luschka, in the early 60's, made a microscopic study of the gland; and since that time, Arnold, Kölliker, and others have written about it.

One thing seems to be sure: that the carotid body is not invariably present. In fact, it is frequently absent. Funke¹ points out that it is enclosed in a fibrous capsule, and that a fibrous band comes from the capsule and divides the body into two parts, other bands from the capsule separating each half into lobules. This fibrous tissue contains a multitude of blood-vessels. Funke further points out that the lobules contain cell-collections without definite arrangement, that only rarely do they resemble the structure of a gland, and that in all parts of these lobules blood-vessels are demonstrable. The same observer believes that the lobules result from proliferation of the endothelial cells of the blood-vessels.

To-day, we should describe the carotid body as a structure placed in the bifurcation of the common carotid artery; to the inner side of this vessel, on a lower level than the bifurcation;

* Read before the Philadelphia Academy of Surgery, May 7, 1906.

¹ *Am. Med.* July 16, 1904.

or on the posterior surface of either the external or the internal carotid. It probably always takes origin from the sheath of the internal carotid. In human beings, it is frequently absent. At least it is frequently absent in those beyond puberty. It is encapsuled in fibrous tissue, is fastened to the sheath of the internal carotid, and the gland with its capsule is embedded in a considerable amount of fat. In shape, it is oval; in color, reddish brown. Its size when not enlarged is about that of a grain of corn. The septa from the capsule divide the organ into follicles, or cell-balls; and these cell-balls are composed of numbers of endothelial cells and capillary blood-vessels. A small branch, several branches, or many branches from the carotid pass into the carotid body; and the carotid plexus of the sympathetic nerve is in very close relation with the body. This structure has been studied, of late, by John Funke, Paltauf, Reclus, Marchand, and others. Its function is unknown.

Occasionally tumors arise in this structure; and Dr. Funke, in the previously-quoted article, has collected fifteen cases. In his series, it is shown that the tumors may occur in adolescents or in adults, and in either sex. He quotes the observation of Heinleth that the carotid body undergoes development until puberty, when it ought to atrophy; but that if it fails to atrophy, but continues to grow, a tumor forms. Such a tumor grows very slowly, requiring years to reach any considerable size, and never becoming very large. Sooner or later, however, rapid growth is liable to begin; and it is usually only after years of growth, and when this sudden rapidity of growth has alarmed the patient, that a surgeon is called in.

Early in the case the growth is entirely free from pain, but in the later stages there may be pain in the tumor, pain radiating into the ear, dysphagia, and—as has been pointed out—perhaps pupillary contraction of the same side and facial vasomotor disturbance. In a large majority of the reported cases, there has been distinct transmitted pulsation in the tumor. The skin is movable over the growth; the tumor may be moved from side to side, but not up and down; and there is usually a systolic murmur over the tumor.

I have recently had, in the Jefferson College Hospital, a case of this rare and interesting trouble, and a diagnosis was made before operation. The record of the case is as follows:

The man was 52 years of age. Over twenty years ago he noticed a very small lump on the right side of his neck. He said that when he first found it this lump was not larger than a grain of corn. During many years it slowly but certainly increased in size. A few months ago it began to grow rapidly, and within less than a year of rapid growth it attained the size of a small egg of a hen. He also began to have some difficulty in swallowing, had attacks of redness of that side of the face, and occasionally suffered from pricking pain in and around the tumor. The rapid growth alarmed him, and he decided to consult a surgeon.

An examination showed the tumor to be in the superior carotid triangle, having its lower border on a level with the upper margin of the thyroid cartilage, and its upper border passing to about the level of the angle of the jaw. The external jugular vein was distinctly visible passing over it. The skin was freely movable over the tumor; and the tumor itself was movable from side to side as though on a hinge, but was not movable from above downward or from below upward. The growth was not tender on handling, but was the seat of very marked pulsation, which investigation demonstrated not to be expansile pulsation, but a lifting of the growth by the pulse of the carotid. The tumor was hard, but somewhat elastic, being, however, softer at some points than at others. It was smooth, but apparently lobulated on the surface. On listening with the stethoscope, a systolic murmur could be made out when the stethoscope was pressed firmly upon it; but this was not more manifest than it was on the carotid artery itself, when the same maneuver was executed.

It was evident that this tumor was not an aneurysm, from its long history, from its hardness, from the absence of genuine bruit and expansile pulsation, and from the fact that pressure on the artery did not cause the mass to diminish in size. It was not a cyst, because it was evidently a solid body. The question of a misplaced fragment of thyroid tissue was considered; but the density, the history, and the vascular phenomena led to the rejection of this idea. It was too hard and too deep for a fatty tumor. Its movability, its long history, and the phenomena

of pulsation were against sarcoma; and the long history, without any change in consistency and without the involvement of the overlying parts, was considered to rule out lymphatic glandular trouble.

I advised operation, on account of the rapid growth then taking place and the apparently inevitable disaster, if this rapid growth were permitted to continue unchecked.

After having exposed the tumor by an incision at the anterior margin of the sternocleidomastoid, and while endeavoring to free it, I was greatly embarrassed by the profuse bleeding. The fatty tissue about the tumor and the capsule of the tumor oozed continuously from numberless places. The bleeding was both arterial and venous. Forty ligatures failed to arrest the bleeding.

After exposing the tumor thoroughly, the growth was found to be in and around the angle of bifurcation of the common carotid; and it embraced the vessels so completely that it was out of the question to free the growth from them as I had hoped to do and as was done in 3 reported cases. It was equally impossible to abandon the operation, because the persistent hæmorrhage barred such a road of retreat. Consequently, the operation was proceeded with.

The common carotid artery was tied with two ligatures below the growth, and was divided between the ligatures. The distal stump of the divided artery was grasped with forceps, and used as a handle in lifting the tumor while the growth was being separated. The tumor, with the beginnings of the internal and external carotid arteries, was freed from its attachments. During this separation the internal jugular vein was badly torn; and it was necessary to ligate it. When the portion of the external carotid artery above the tumor was reached this vessel was tied and divided. Between the upper border of the tumor and the base of the skull there was barely room to ligate the internal carotid; it was with great difficulty that it was ligated and divided, and I barely escaped the accident met with by Mikulicz, who found the tumor had entered the bony foramen and was obliged to cut away bone to stop bleeding. The wound was closed with drainage.

The man had lost much blood and was considerably shocked. He reacted but slowly from the anæsthetic. Eight hours after the operation he developed a weakness just short of complete paraly-



FIG. 1.—Case XXVI.

sis of the left arm and leg, the face escaping. He was also found to have a very low and extremely hoarse voice. The day after the operation, the voice continuing low and hoarse, the throat was examined; and relaxation and œdema of the right vocal cord were observed by Dr. J. Leslie Davis to exist. These conditions were due to paralysis of the cricothyroid muscle from injury of the superior laryngeal nerve.

For many days there was a copious flow of mucus from the larynx and the bronchi; and, owing to the anæsthesia of the mucous membrane, the patient had great difficulty in expelling this mucus. For some time there was considerable difficulty in swallowing, probably also due to injury of the superior laryngeal nerve, which, it will be remembered, also goes to the inferior constrictor of the pharynx. For the first few days after the operation there was a copious flow of lymph from the wound, showing that large lymphatic vessels had been divided. This ceased about the end of the first week.

On the eighth day after operation complete hemiplegia suddenly developed. The left arm and leg were completely paralyzed; the face was much drawn; and the man was dull, drowsy, and sometimes stuporous, but never unconscious. It was the opinion of Dr. Alfred Gordon that this attack was due to embolism, in all probability in the internal capsule; and the first and milder attack was thought to have been due to thrombosis in the cortical vessels.

The day after the onset of the hemiplegia, the man was found to be suffering severely with dyspnœa and repeated choking fits, in some of which it seemed that he must strangle. Great quantities of mucus passed into the throat, and there was the greatest possible difficulty in ejecting it. Examination of the left lung, made by Dr. John C. DaCosta, Jr., developed the fact that at least half of the lung was in a state of complete collapse, containing no air whatever. The right lung was entirely normal. The patient stated that he had had an exactly similar pulmonary condition a number of months before. This had come on from an unknown cause, and had almost killed him. The atelectasis produced great discomfort for a number of days, but was gradually recovered from; and the lung is now normal, so far as physical signs indicate. It seems probable that the laryngeal anæsthesia was responsible for this condition,

and that either plugs of mucus had passed into the lung and blocked the bronchi, or that some elements from the food had passed the larynx.

Present Condition (8 weeks after the operation).—A marked, but fading, left hemiplegia exists. The man can move the leg, and can stand upon his legs, if he supports himself with a cane or a crutch. He can move the elbow, the shoulder, and the wrist, and can flex the hand; but the extremity is still very weak. He has occasional paroxysms of violent shooting pain in the arm and in the leg. The wound is completely healed and not tender. The voice is hoarse and low, and the right vocal cord is oedematous and relaxed; and Dr. Davis is of the opinion that this is due to injury of the superior laryngeal nerve.

Conclusions.—It is thus seen that the operation of removing a tumor of the carotid body is a very formidable one. The surgeon may have to tie all the carotid arteries; and he may damage a nerve or nerves, with subsequent unfortunate results. The ligation of the common carotid artery is an extremely dangerous procedure; and it is one of the few operations in which the mortality does not seem to have been greatly diminished since the days of Sir Astley Cooper, who did the first successful ligation of the common carotid, in 1808. Mr. Richard Barwell, in his article on Aneurysm in "Ashhurst's International Encyclopedia of Surgery," published in 1889, gives the mortality of 107 cases of ligation of the common carotid for aneurysm as 25.23 per cent. Some more modern authors estimate the death-rate as in the neighborhood of, or over, 30 per cent.; and it is thus seen what a responsibility it is, even at the present time, to tie this vessel.

The danger of death is, however, not the only danger in ligating the common carotid. My case shows that hemiplegia may follow the operation. It has long been known that a considerable percentage of those on whom ligation has been performed suffer subsequently with cerebral symptoms. In some of the cases, these symptoms have been produced by thrombosis; in others, by embolism; and in still others, by cerebral softening. Pilz has pointed out that 32 per cent. of the cases



FIG. 2.—Case XXVI. Section.

in which the common carotid has been ligated exhibit brain symptoms, and that 56 per cent. of the cases that show brain symptoms die. Zimmermann says that in 11 per cent. of the cases there is softening of the brain, and that 26 per cent. of the cases show brain symptoms. There is much greater danger of brain symptoms when the operation is performed on the elderly or middle-aged than when it is done on the young. In older subjects, arterial atheroma may interfere with the distension of certain vessels whose integrity is necessary to bring sufficient blood from the vertebrals, from the other internal carotid, and from the terminations of the external carotids. Failure in a satisfactory restoration of circulation is most liable to occur when profuse bleeding greatly lowers the blood-pressure, as it did in this case. When such cerebral change ensues, it does not necessarily mean death. In fact, it may be recovered from, partially or completely. Usually, however, the condition is permanent and progressive, and finally results in death. In Funke's series of 15 cases of tumor of the carotid body, there were but two deaths; one from bronchopneumonia, and one from secondary hæmorrhage. There may be added to this, Keen's unreported case, which makes three deaths in 16 cases. In Funke's series, there was but one case of hemiplegia. In Dr. Hearn's case, however, which is not recorded in the table as one of hemiplegia, the patient died two months later; and Dr. Hearn tells me that, although he did not see the case, he believes from what he has learned that the man died of cerebral softening. So, out of 13 recoveries in Funke's cases, to which my cases may be added, making 14 recoveries, there were two cases of hemiplegia and one of cerebral softening.

Owing to the great danger in ligating the common carotid, surgeons have sought to avoid it in removing carotid tumors. The reported cases show that almost always all the carotids must be ligated. Albert, in his case, was obliged to ligate only the external carotid, being able to remove the growth from the carotid sheath. In his case the growth recurred within one year. In Heinleth's case and in that of Cuneo, no ligations were necessary, owing to the free separability of the tumor.

Out of Funke's 15 cases, only three are recorded as not requiring ligations of all the carotids; and when Keen's case and mine are added to this list, they make 17 cases, in 14 of which ligation of all the carotids was necessary.

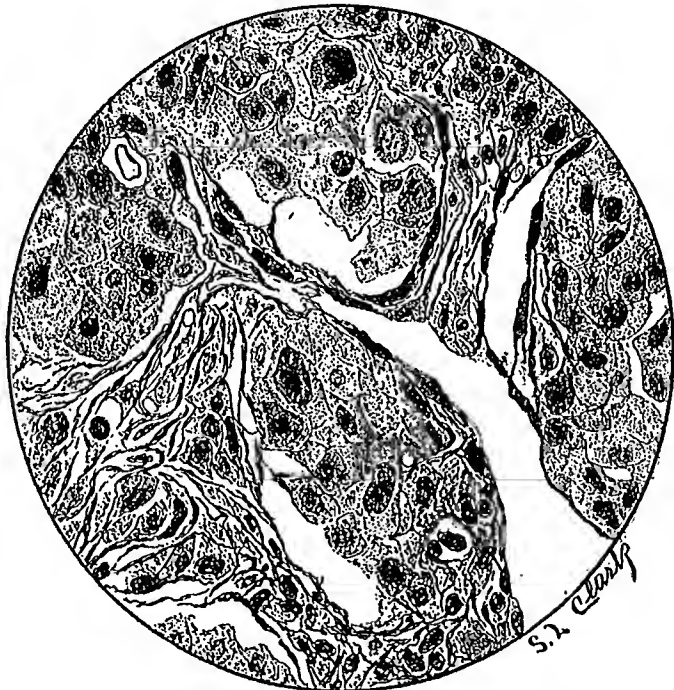
Another danger of the operation is nerve-injury. In my case, the superior laryngeal was injured. In all probability, it is a piece of this nerve that we find incorporated in the specimen. In Funke's 15 cases, there were six cases in which the nerves were injured—the sympathetic, the pneumogastric, the hypoglossal, the facial, or the recurrent laryngeal. Two cases out of the 15 exhibited postoperative paralysis of the vocal cord of the side operated upon.

It is thus evident that the operation of removing a carotid tumor is an extremely dangerous one, and is not to be lightly undertaken. We agree with Reelus that one should not touch these growths, unless they are productive of danger to life. So long as they are merely slowly progressing, they had better be let alone. It is only when they begin to grow rapidly that one should remove them, and then he must, in spite of the danger. In my case, the tumor was infiltrating the surrounding structures, and would unquestionably have killed the man, if allowed to remain. Surgeons must be wide awake to the existence of such growths. Without carefully examining every tumor in this region of the neck, one could easily be led into operating with a light heart for some supposedly trivial condition, and then find oneself suddenly so far advanced in attacking a carotid tumor that retreat would be impossible, and probably all the carotids would have to be tied. The diagnosis is possible in many cases. It was made in several of the cases in Funke's list, and it was made in the case now reported. The pictures exhibit the tumor that I removed, and Dr. Funke's report of the specimens follows:

Macroscopic Description.—The specimen is a lobulated mass, measuring 5 by 5 by 4 cm.; weight 104 gmm. It is dark red in color, encapsulated, distinctly elastic in consistency at some places and flabby at other places. The mass is irregular; it is composed of three large nodules, each being 2.5 cm. in one diameter and 2 cm. in the other. The smaller

nodules present do not attain a diameter of 0.5 cm.; they are especially seen on the anterior surface. This surface contains many depressions varying from 0.5 to 1 cm. in depth; these depressions are incident to the pulling of the capsule into the tumor substance. The lacerated tissue present adds to the irregularity of the anterior surface. The posterior surface is less irregular; it is lobulated, however, and the nodules are more conspicuous here than upon the anterior aspect. Laceration and

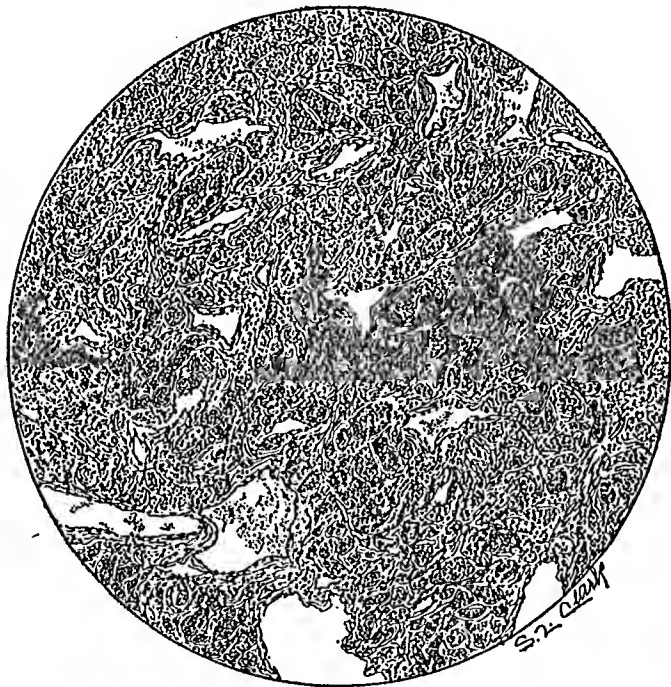
FIG. 3.



fragmentation of the capsule is marked. Lying upon this surface, not more than 2 cm. from the margin, is a greyish-pink cord-like piece of 'issue apparently made up of smaller cords; the consistency and the architecture of this structure resemble a nerve. It is not firmly attached to the tumor-mass. Lying upon this surface, but only near one end, is a large vessel which from its general structure appears like the common carotid artery and which contains, 0.5 cm. from the free margin, a liga-

ture. One centimeter above the ligature the vessel divides; one branch curves slightly toward what was described as the anterior surface and then tunnels the mass between two of the nodules described. Only 0.5 em. of this branch is visible, but upon dissection it is found to traverse the mass nearly parallel with the anterior surface and but 0.7 em. from it. The other vessel curves slightly backwards and then tunnels the mass near the opposite side of the specimen, but runs parallel and very close to the

FIG. 4.



posterior surface. Both vessels are easily identified at what is presumed to be the superior portion of the tumor, and both vessels as well as the common carotid artery are firmly attached to the tumor mass. The first vessel described in all probability was the external carotid, since it gave off a small branch near its point of severance.

Dissection showed that the three larger nodules mentioned are firmly united at a point posterior to the bifurcation of the vessel mentioned. The one nodule is united to the other two at this point and along the

entire margin of but one nodule by means of a pedicle; the internal carotid passes between these nodules and is anterior to the pedicle. The other two nodules are for the most part situated in the fork formed by the branching vessel. Dissection also reveals that the smaller nodules are produced by the septa which penetrate from the capsule into the underlying tumor mass.

The cut surface has a lobulated appearance; it is granular, reddish-brown in color, but traversed by greyish bands; some of these bands are dense and comparatively broad. The cut surface as well as the capsule contains many small opened-mouthed blood-vessels; so numerous are they the surface has a porous appearance.

Portions of the tumor were fixed in Zenker's fluid and the remainder was preserved in Kaiserling's fluid. Sections were made and stained with hematoxylin and Van Gieson's method for connective tissue, by Mallory's reticulum stain and with polychrome-methylene blue.

Histology.—One margin of the sections is covered by a dense capsule composed by wavy fibrous connective tissue, in which are few lymphoid and spindle-shaped cells and few strands of elastica, together with many blood-vessels. From the capsule fibrous septa penetrate the underlying tumor-mass and divide it into lobules, which are again divided into alveoli. The fibrous septa are very broad and are found in cross and in longitudinal sections; they contain few lymphoid and spindle-shaped cells and large and small blood-vessels. Many of the last-named structures contain erythrocytes, and possess well-formed and thick walls.

The walls of the alveoli are in some instances formed by delicate connective-tissue strands, evidently constituents of the septa already mentioned; the greater number, however, are formed by delicate capillaries, branches of the vessels found in the septa. Occasionally these capillaries are composed of a single layer of endothelial cells; in other instances the endothelial lining is supported by a few strands of fibrous connective tissue. The alveoli are fairly uniform in size and very difficult to outline in many places, owing to the number of contained cells. The cells in the alveoli vary somewhat in size, ranging from 15 to 25 microns in diameter; they are irregular in outline, many are polyhedral and few are oval. The protoplasm contains no cell membrane; it is finely granular and acidophilic. The nuclei are comparatively large and intensely basophilic. The nuclear membrane is conspicuous. Occasionally few red blood-cells are found among the tumor-cells. In not a few alveoli the tumor-cells show degenerative changes. Few chromaffin cells are present.

Diagnosis.—*Endothelioma*; this is the type of tumor to which most writers on the neoplasms of the carotid gland apply the term "perithelioma." From the fact that the growth has invaded the vessels and the surrounding tissues it should be looked upon as malignant.